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ZORA URL: <https://doi.org/10.5167/uzh-148510>

Journal Article

Published Version

Originally published at:

Farshad, Mazda; Winkler, E; Betz, M (2017). Anterior Surgical Treatment Of Scoliosis In A Patient With Loeys Dietz Syndrome. Journal of American Academy of Orthopaedic Surgeons, 1(7):1-5.

Anterior Surgical Treatment of Scoliosis in a Patient With Loeys-Dietz Syndrome

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None of the following authors or any immediate family member has received anything of value from or has stock or stock options held in a commercial company or institution related directly or indirectly to the subject of this article: Dr. Farshad, Ms. Winkler, and Dr. Betz.

JAAOS Glob Res Rev 2017;1:e039

DOI: 10.5435/
JAAOSGlobal-D-17-00039

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Abstract

Loeys-Dietz syndrome (LDS) is an autosomal dominant disorder affecting the connective tissue, resulting in laxity, and can be challenging if surgical treatment is needed. Literature concerning scoliosis and its treatment in LDS is limited. This is a report of scoliosis surgery in a 12-year-old girl with LDS. She underwent anterior instrumented spinal fusion of the segments T12 to L3 because of a left-sided thoracolumbar scoliosis of 42° with dysplastic pedicles. The scoliosis was reduced by 40%, from 42° to 25°, resulting in a satisfactory clinical outcome with a minimal amount of surgical invasion and lack of complications. A minimal approach and anterior instrumented spinal fusion surgery can be considered a valuable surgical treatment alternative for scoliosis in patients with LDS, avoiding the placement of pedicle screws in dysplastic pedicles and using the abnormal laxity of the connective tissue as an advantage.

Loeys-Dietz syndrome (LDS) is a rare autosomal dominant disorder affecting the connective tissue.¹ Manifestations of LDS include aortic aneurysmal disease, vascular fragility, congenital heart problems, and thin and translucent skin, as well as skeletal abnormalities, such as pectus deformity, osteoarthritis, and spinal deformities.¹ Cervical malformations and anomalies are important features of LDS and can result in cervical instability.² Because LDS is extremely rare, reports on treatment of scoliosis in such patients are limited.

In regard to the spine, both Marfan syndrome (MFS) and Ehlers-Danlos syndrome (EDS) seem to have similarities to LDS, so that the knowledge on their treatment

strategies can potentially be used in the management of LDS. Here, we report a surgical technique of scoliosis correction applied to a patient with severe laxity, in which the laxity was used to technical advantage to enforce the surgical treatment strategy.

Case Report

Patient's History

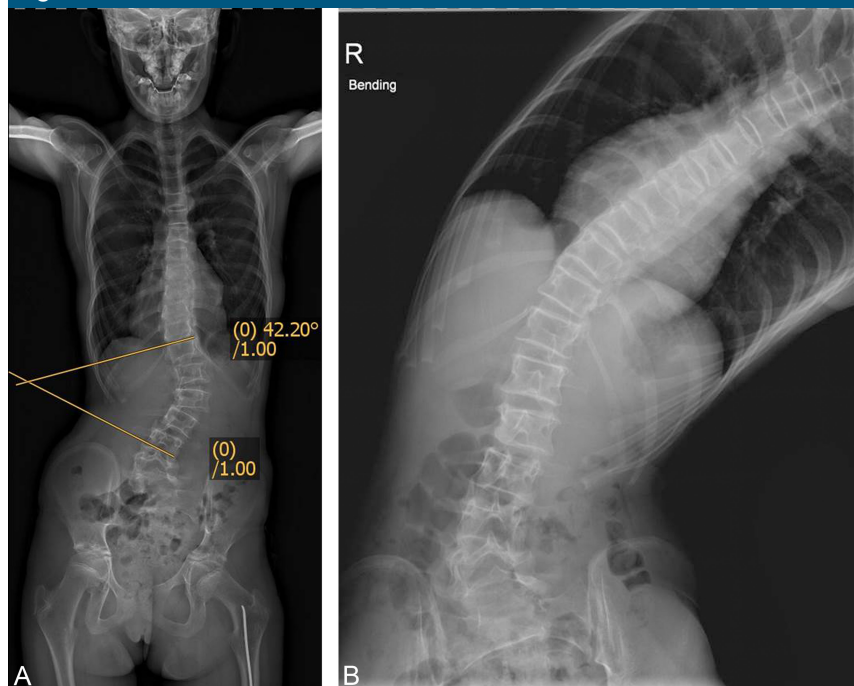
A 12-year-old girl was referred for evaluation of a left-sided thoracolumbar scoliosis of 42°. She was diagnosed with LDS on the basis of clinical findings and genetic testing. The scoliosis had been treated with a corset, which at first was worn only

Figure 1



Photograph showing the patient from the back. Note the tall stature with disproportionately long, slender limbs and scoliosis.

Figure 2



(A) Preoperative AP radiograph showing a curve of 42°. (B) Supine side bending to the left side showed good flexibility of the lumbar curve.

at night, but later, with progression of the curve, also during the day.

Documented cardiovascular findings included mild tricuspidal and mitral valve prolapse, slight dilatation of the annulus fibrosus, an aortic sinus, and a microaneurysm with a dilatation of 1.7 mm in the P1 segment of the posterior cerebral artery. Angiotensin-II antagonist (losartan) therapy had already been initiated to keep blood pressure levels low and decrease transforming growth factor beta signaling.

Clinical Examination

On initial examination, the patient was of tall stature and presented with a marfanoid habitus, arachnodactyly, camptodactyly, joint hyperlaxity, and left-sided thoracolumbar scoliosis. The patient had a cleft palate and thin, translucent skin.

The scoliosis was measured as a 1.5-cm truncus shift to the left, with no shoulder obliquity but asymmetrical

waist triangles (Figure 1). The forward-bending test showed a prominent lumbar hump on the left side at L1-2. A pelvic obliquity on the left side and a 2-cm longer right leg were also noted. Skeletal maturity was classified as Risser sign 0, indicating significant remaining potential for growth and curve progression.

Neurologic examination documented normal sensation and overall normal motor activity of the lower and upper extremities.

Radiologic Examinations

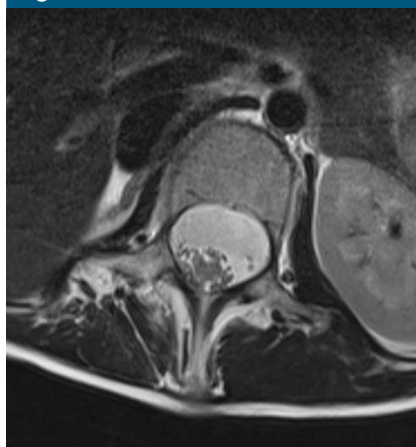
On standing AP whole-spine radiographs, two regional curves were identified: a left-sided lumbar curve (T12-L4; apex, L2) that progressed gradually from 34° to 42° during the period of 2 years, and a compensatory right-sided thoracic curve (T8-T12; apex, T10), with a Cobb angle of 19° (Figure 2, A). Supine side bending to the left side showed good flexibility of the lumbar curve,

with a curve correction to almost 0° (Figure 2, B).

Treatment

With a progressive scoliotic curve of 42° in the skeletally immature (Risser sign 0) patient, correction of the curve was indicated. Curve flexibility allowed the possibility of an anterior approach, thereby avoiding the challenges of pedicle malformation and significant dural ectasia (Figure 3).

Perioperative management included avoidance of high peaks in blood pressure to minimize the risk of arterial dissection. This was achieved with sufficient pain management by means of fentanyl (patient-controlled) analgesia, paracetamol, and metamizole. Heart functions and valve insufficiency were not relevant enough to induce specific anesthetic precautions. Losartan was replaced with nitroprusside perioperatively because of its fast-acting properties and high vasodilating efficacy.

Figure 3

Magnetic resonance image showing dysplastic pedicles and dural ectasia.

A retropleural approach was performed without a pleurotomy to avoid the need of a chest tube. Discectomy of the intervertebral disks T12-L1, L1-L2, and L2-L3 was performed. Instrumentation allowed a moderate hold, despite modest bone quality. Somatosensory and motor evoked potentials showed stable and steady values throughout the procedure. Surgical time was 3.5 hours, and blood loss was 400 mL.

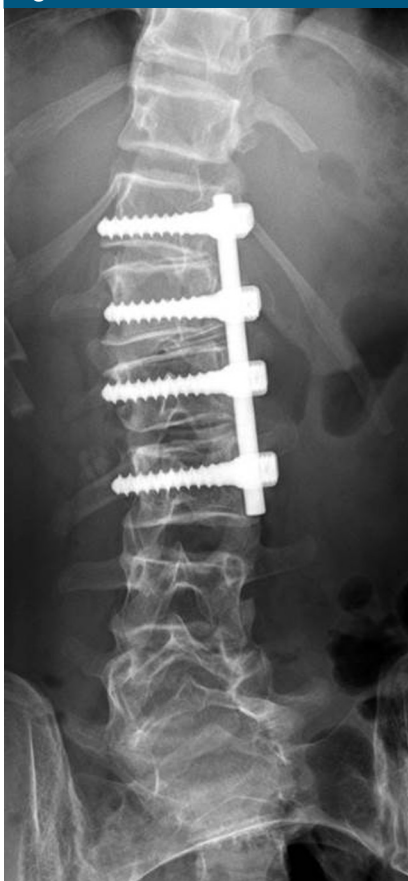
Postoperative standing whole-spine radiographs demonstrated a satisfactory correction of the curve (Figure 4).

A brace was applied for 6 weeks following surgery, and the patient was instructed to avoid flexion/extension or rotation of the spine.

At 12 months of follow-up, the patient was satisfied and showed no neurological deficits (Figure 5). Radiographic images showed a phenomenon of compensation at the segment L3-4 (Figure 6).

Discussion

This is the first report of a minimal approach and anterior instrumented

Figure 4

Postoperative radiographs showing correction of the scoliosis curve.

spinal fusion surgery for the treatment of scoliosis in a patient with LDS, thus avoiding the placement of pedicle screws in dysplastic pedicles and using the abnormal laxity of the connective tissue to advantage. The current literature concerning scoliosis treatment in LDS is limited.

Patients with syndromic scoliosis, such as in MFS, EDS, or LDS, tend to be at a higher risk of perioperative complications than are patients with idiopathic scoliosis. Complications include dural tears/cerebrospinal fluid leakage, wound infections, respiratory issues, neurologic compromises, hardware failure, greater-than-average estimated blood loss, and pseudarthrosis.³⁻⁵

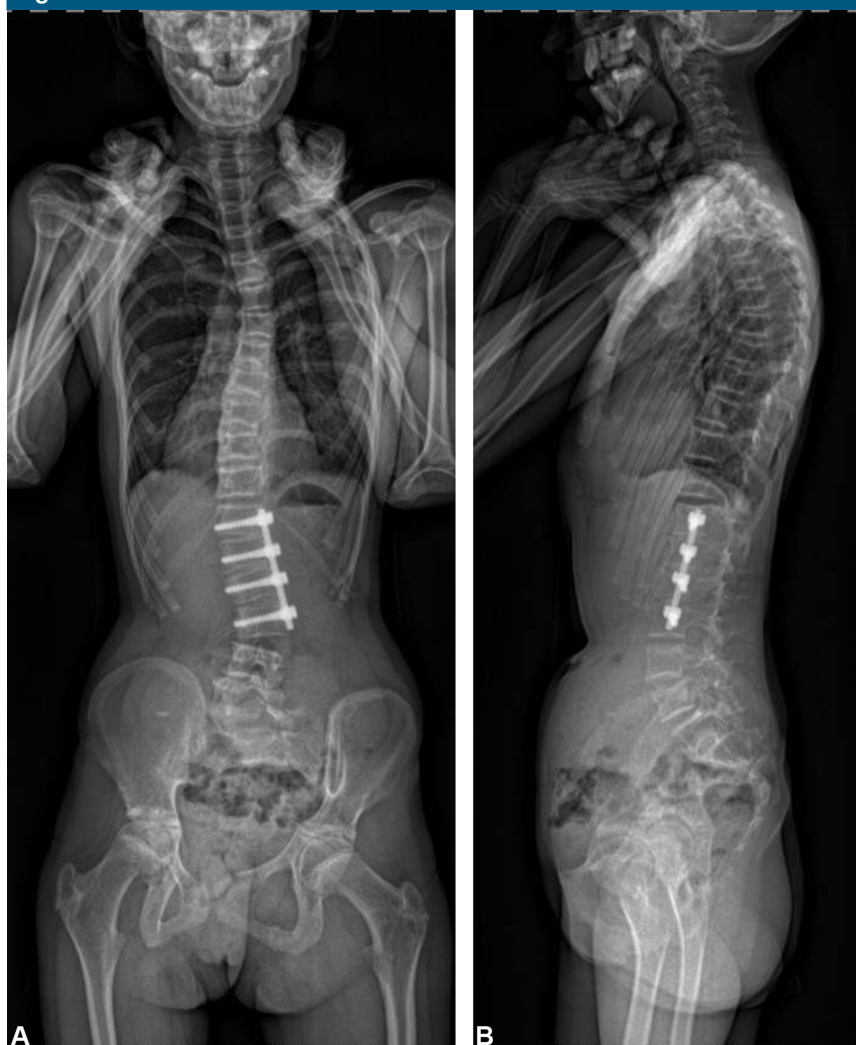
Figure 5

Postoperative photograph at 12 months of follow-up.

The most recent published study, conducted by Bressner et al⁶ discussing scoliosis in patients with LDS and comparing different treatment options, showed that bracing failed in 11 of 15 patients. Contrarily, the same group noted high rates of complications in patients with LDS who underwent growing rod surgery or spinal fusion. In 11 of 24 surgeries, the blood loss was >20% of the estimated total blood volume, which the authors assumed to be linked to the fragility of the vessels. They further observed growing rod fractures in 2 of 3 patients; cerebrospinal fluid leakage in 24 cases, which they suggested was partly due to the high prevalence of dural ectasia in these patients; and patients requiring extension of fixation to additional vertebral levels or to the pelvis. However, no revisions due to failure of fixation of screws or hooks were needed.⁶ Controversies exist concerning the surgical approach in syndromic scoliosis.

Li et al⁷ showed that posterior-only surgery with instrumented fixation

Figure 6



AP (A) and lateral (B) radiographs made 12 months after surgery showing an acceptable curve correction.

and fusion is effective and safe for the treatment of scoliosis associated with MFS. Jasiewicz et al⁸ presented a study of 11 patients with EDS. Six patients were treated by posterior fusion and five patients by a combined anterior and posterior fusion. No intraoperative complications were reported, but reoperations were needed because of increasing sagittal imbalance or instrumentation failure in four cases. Akpinar et al⁹ presented five patients with EDS undergoing surgical scoliosis correction. In four patients, anterior discectomy and fusion as well as posterior

instrumentation were performed. The fifth patient underwent posterior surgery alone. Complications during anterior lumbar surgery included avulsion of segmental arteries from the lower aorta, as well as rupture of the iliac vein and artery due to blunt dissection. The authors suggested avoiding blunt dissection for exploration of the vessels, carefully isolating and ligating segmental arteries as far away from the aorta as possible, and minimizing discectomies. Yang et al¹⁰ reported of three patients with EDS who had excessive bleeding during anterior surgery. They warned

that complications during anterior surgery in patients with EDS occur frequently.

Here, we presented the first report of anterior instrumented spinal fusion surgery for a patient with LDS, thereby avoiding the insertion of pedicle screws into dysplastic pedicles and the potential complications associated with dural ectasia. We also used the abnormal laxity of the connective tissue in LDS as a technical advantage. In our patient, no vascular complications occurred during or around the time of surgery. At 12 months of follow-up, radiographic images showed an acceptable correction, and the patient was satisfied. Hyper-compensation of the caudal adjacent segment is another result of the extensive hyperlaxity in LDS.

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